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SMART PEDI UPDATE

Pedismart Newsletter
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Pedi smart News Letter
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Dear Doctor,
Greetings from Micro Labs Limited. We are happy to bring you **"PEDI SMART"** which contain latest and interesting news in the field of Paediatrics.

This issue contains:

1. **A study on early echocardiographic signs of cardiovascular affection in pediatric familial hypercholesterolemia**
2. **A study on carboxyhemoglobin, a biomarker of late-onset sepsis in preterm infants**
3. **A study to evaluate the associated risk of bed sharing among mother-infant dyads**
4. **A rare case study on Discoid lupus erythematosus in an 8-year-old girl**

Should you require any specific article or medical information, please feel free to contact us at medicalservices@microlabs.in

Looking forward to your valuable feedback

Best Regards,

Medical Team, Micro Labs Limited

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SMART PEDI UPDATE

A study on early echocardiographic signs of cardiovascular affection in pediatric familial hypercholesterolemia

Introduction –

Familial hypercholesterolemia (FH) is an autosomal dominant genetic disorder which is characterized by elevated levels of low density lipoprotein cholesterol (LDL), which can cause atherosclerosis, cardiovascular problems, and early mortality. There are two different forms of FH: homozygous and heterozygous variations[1]. Plasma LDL-C levels that have homozygotes of FH (HoFH) may be four times higher than average levels and at least twice as high as those of heterozygous FH (HeFH). Familial hypercholesterolemia causes a delayed clearance of plasma LDL2 because of anomalies in the genes implicated in the LDL receptor-mediated pathway[2]. This study aimed to determine the effect of hypercholesterolemia on the cardiovascular system with homozygous FH using echocardiographic parameters such as tissue Doppler imaging (TDI) and 2-dimensional speckle-tracking echocardiography (2D-STE) in pediatric patients.

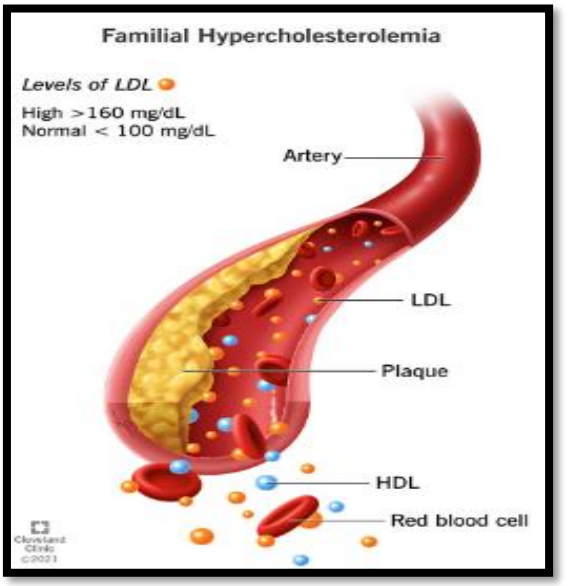

Fig.1- Familial hypercholesterolemia

Methods –

This is a case control study that included 21 children with familial hypercholesterolemia and 25 healthy controls. Clinical and laboratory data were collected, and echocardiography was performed for both groups of pediatric patients. Modern echocardiographic techniques such as TDI and 2D- STE was performed for all cases. Statistical analysis was performed and for comparison between groups was done by using non-parametric Mann-Whitney U test for non-normally distributed numeric data, the Student T test for normally distributed numeric data, and the chi-square test for categorical variables. The correlation between variables was identified using Pearson and Spearman correlation coefficients.

Results –

According to the findings, the patient group had greater systolic blood pressure than the control group, and all of the patients had xanthomas and a favourable family history. The majority of FH patients (85.7%) had mild aortic insufficiency, according to conventional echocardiography (Fig. 1 A). The left ventricle was considerably hypertrophied in 2D pictures and M-mode echocardiography (Fig. 1c), and the right and left major coronary arteries were more dilated in patients than in controls (Fig. 1B). The patient group had lower fractional shortening (FS) and ejection fraction (EF) values, but a larger left atrium (LA). In the patient group, mitral E/E′ was greater and there was moderate diastolic dysfunction in the LV. When compared to the control group, the abdominal aorta circumferential strain and ascending aorta M-mode-derived strain

were lower in the patients, indicating enhanced aortic stiffness in the patients' group. Mitral E/E′ was correlated positively with the LDL cholesterol level and aortic elastance.

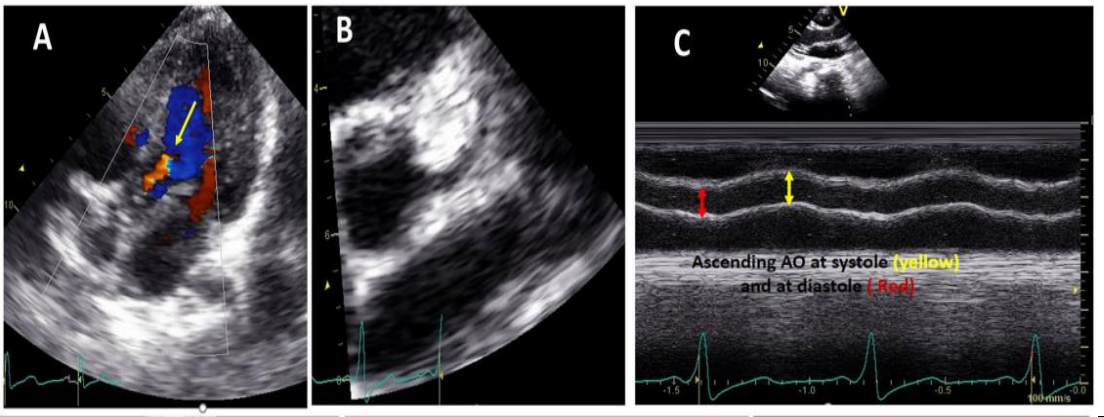

Fig.2 (A) - Color Doppler showing mild aortic insufficiency. (B) - Left coronary artery showing left main coronary artery ectasia. (C) – M-mode measuring systolic (yellow arrows) and diastolic (red arrows) aortic dimensions.

Discussion –

Current study states that due to increase in intimal endothelium thickness caused by hypercholesteremia, the blood vessels become stiffer and the blood pressure rises. This may provide an explanation for the patient group's blood pressure readings' rising systolic values. FH can affect heart valves, especially the aortic valve, leading to aortic valvopathy. Of the patients, 85% had an insufficient aortic valve, whereas 14% had an enlarged aortic valve. With median values in the patient group of 34% and 64%, respectively, which are still within the normal ranges for the paediatric age group, M-mode LV systolic parameters, FS and EF were considerably lower in patients compared to those in the control group. Aortic distensibility and aortic change area, which were lower in the FH group than in the control group, showed a statistically significant difference. A study by de Boer LM et al., showed that in patients with familial hypercholesterolemia, lipoprotein concentrations contribute significantly to arterial wall thickening leading to risk factor for early atherosclerosis[3]. According to the study by Pederiva et al., the risk factors for positive family history of premature cardiovascular disease (pCVD) are Children/adolescents with FH and Lipoprotein > 30 mg/dl where more likely[4].

Conclusion –

Aortic valve stenosis, coronary artery enlargement, left ventricular (LV) diastolic dysfunction, LV hypertrophy, and increased aortic stiffness are early cardiovascular indicators in children with homozygous FH. These signs could appear before the LV systolic function declines.

Homozygous familial hypercholesterolemia is associated with cardiovascular risk factor in pediatric patients

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A study on carboxyhemoglobin, a biomarker of late-onset sepsis in preterm infants

Introduction –

Carbon monoxide (CO) is formed naturally and endogenously through various enzymatic and non-enzymatic pathways. Heme oxygenase (HO), which catabolizes heme to CO, iron, and biliverdin, is responsible for producing around 85% of CO. Carboxyhemoglobin (COHb) is formed when CO and haemoglobin link together instead of oxygen. On the other hand, oxidative stress and inflammation caused an increase in HO, and conditions associated with oxidative stress, primarily sepsis and shock in both adult and paediatric populations, have been reported to have high levels of COHb[1]. Due to the pathogen microorganisms, late-onset sepsis (LOS) usually occurs with the onset of symptoms 72 hours after birth. The risk of infection is inversely correlated with gestational age and birth weight. [2]. This study aimed to assess the correlation between COHb levels and the risk for LOS development.

Methods –

In this retrospective observational study inborn children with gestational ages under 30 weeks who developed LOS were included. There were 100 preterm infants in total, 50 of whom were in the LOS group and 50 of whom were not. A blood gas analyser was used to measure the amounts of COHb in blood samples. COHb levels were assessed on the day that the first episode of LOS was diagnosed, 3, 2, and 1 days earlier, as well as 1 and 4 days afterwards. The total neutrophil count, C-reactive protein, and procalcitonin tests were used to make the diagnosis of sepsis, which ws then supported by the discovery of at least one positive blood or alcohol culture. The primary aim of the study was to determine whether COHb levels and the likelihood of LOS formation would be correlated. In order to rule out discrepancies, the secondary endpoint compared the COHb levels of infants who experienced Gram-positive sepsis with those who developed Gram-negative sepsis. A probable association between the risk of LOS and characteristics that, in a univariate analysis, differed across the groups was examined using logistic regression analyses. The predictive usefulness of the COHb level,

which was measured 2 days prior to the onset of LOS, was examined using ROC (receiver operating characteristic) curve analysis.

Results –

Infants in the LOS group developed LOS at 16± 12 days of life. COHb levels measured 2 days before the diagnosis of LOS which were higher in the LOS than in the control group. Logistic regression analysis showed that a higher level of COHb 2 days before the diagnosis of LOS increased the risk for developing LOS, as well as the need for mechanical ventilation. According to ROC analysis, COHb measured two days before to the diagnosis strongly indicates LOS, with COHb=1.55% being the best predictive cut-off point with a 70% sensitivity and specificity. (Figure.1).

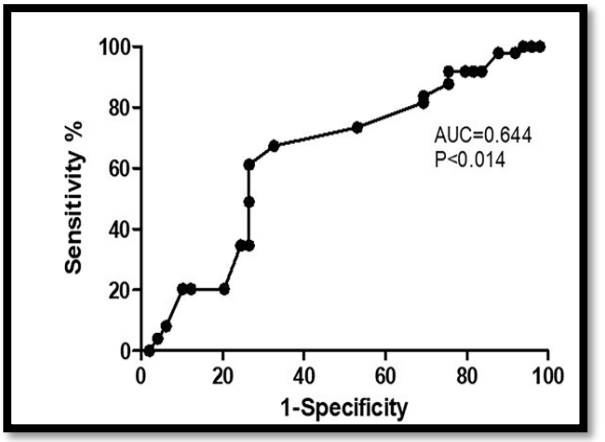

Fig.1- ROC curve analysis for COHb blood levels measured 2 days before the diagnosis of late-onset sepsis.

Discussion –

In this current study, when measured in the days just before its onset of sepsis, COHb blood levels in extremely preterm new-borns can predict the development of LOS. Before to the diagnosis of LOS, COHb blood levels were observed to have significantly increased, and 2 days before to the diagnosis of LOS, COHb levels in infants in the study group were greater than in the control group. Regression analysis showed an independent correlation between COHb levels assessed two days prior to the diagnosis of LOS and the risk for LOS. Additionally, the best LOS prediction threshold is a COHb level of 1.55%, obtained two days before to the diagnosis of LOS. A study by Guney Varal et al., showed that COHb readings of the LOS group significantly increased during the time of the LOS episode. ROC analysis showed COHb level of 1.35%, Sensitivity level was 56.07 and specificity level was 90. The study therefore implies that the COHb levels can be easily determined at the patient's bedside from small amount of blood, which may contribute to the early detection of LOS[3].

Conclusion – This study concludes that, elevated COHb levels may help to predict the diagnosis of LOS in very premature infants. Additionally, this may serve as a biomarker for neonatologists to monitor and detect sepsis in high-risk patients earlier.

COHb levels help for early diagnosis of LOS in very preterm infants

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A study to evaluate the associated risk of bed sharing among mother-infant dyads

Introduction –

Co-sleeping is the activity of a child sharing a bed with a parent(s), or sleeping in the same room but on a different sleep surface[1]. The American Academy of Paediatrics (AAP) estimates that each year in the United States, about 3500 new-borns still pass away from sleep-related causes. For the purpose of enhancing baby safety while sleeping, the AAP updated its safe sleep recommendations in 2022. To reduce baby sleep-related mortality, the revised guidelines included using sleep surfaces and locations, dressing infants for sleep, feeding them with human milk, and avoiding exposure to tobacco, alcohol, and drugs. The AAP remains to be clear that it never advises bed sharing.[2]. This study aimed to test the hypothesis that sleep difficulty at 1 month of age would be associated with bed sharing in the first 12 months, irrespective of social determinants of health (SDOH).

Methods -

This prospective, longitudinal cohort study analysed how breastfeeding mother-infant dyads slept from conception to 12 months of age. Infants born within seven days of birth and singletons delivered at term (37–42 weeks' gestation) were included. 191 couples in total completed the poll. The Brief Infant Sleep Questionnaire (BISQ) was administered to dyads who completed the study and separated into two groups: those who shared a bed (n = 38) and those who did not share a bed (n = 153) at 1, 4, 6, and 12 months. Total time asleep at night (hours), total time awake at night (hours), and time required to put infant to sleep (hours) were measured. Primary outcome was bed sharing at any point in the first 12 months following birth.

Results –

According to the findings, babies who continued to share a bed at one month of age slept only marginally fewer at night (7.1 ±1.7 hours) and required longer to fall asleep (0.7±0.6 hours) than babies who did not (8.3 ±1.5 hours and 0.5±0.5 hours, respectively). In the first months, babies (n = 38) who shared a bed slept less hours at night than their counterparts who did not (Figure 1). While controlling for family income and mother education level, a binomial regression analysis found that bed sharing was influenced by the total number of hours spent sleeping at night and the total amount of time needed to put a new-born to sleep.

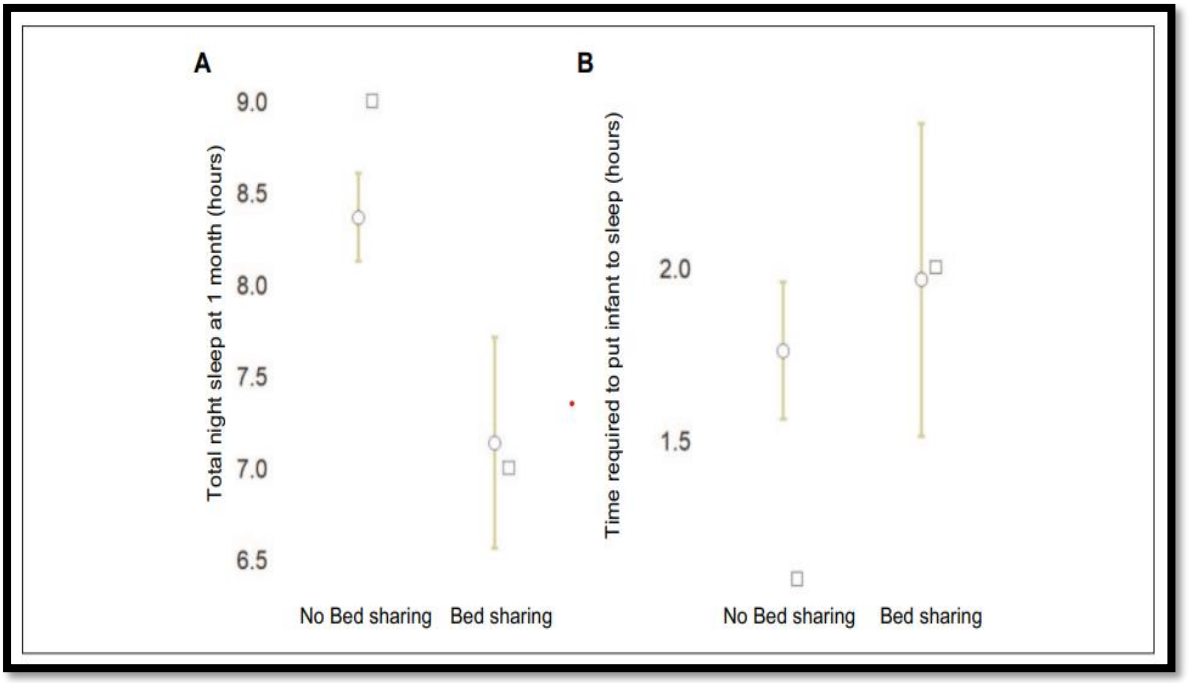

Fig.1 - Whisker plots showing the mean (circle), median (square), and standard deviation (yellow lines) for 2 parameters of sleep difficulty

Discussion –

According to this study, infants that have longer sleep latency and shorter sleep duration at one month are more likely to share a bed throughout the first twelve months. Due to the presence of bulky bedding, maternal smoking, and maternal alcohol use, bed sharing is known to increase the risk of sleep-related new-born mortality in children less than 12 weeks. Early well-child visits to the primary care office, such as the new-born, one-month, and two-month visits, can help parents get informed about sleep issues and the dangers of bed sharing[2]. According to study by Bartick M et al., in some hazardous circumstances, bed sharing is linked to an increased risk of sleep-related death. Sofa sharing, sleeping on a chair, exposing infants to cigarettes, sleeping next to an intoxicated adult, and sharing a bed with a low-birthweight or premature baby are all dangerous situations[3].

Conclusion –

Study draws the conclusion that increased rates of bed sharing, a known risk factor for sleep-related infant death, are associated with sleep issues related to increased sleep latency and decrease sleep duration.

Infant age less than 12 weeks, presence of bulky bedding, maternal smoking, and maternal alcohol use were some of the risk factor for infant's death.

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A rare case study on Discoid lupus erythematosus in an 8-year-old girl

Introduction –

Discoid lupus erythematosus (DLE) is a chronic cutaneous lupus erythematosus (CCLE), which is part of the spectrum of cutaneous lupus erythematosus (CLE)[1]. DLE is an inflammatory skin condition that typically strikes in adolescence or early adulthood. Approximately 5-7% of DLE patients begin experiencing symptoms before the age of 16 [2]. The cause of DLE is still unknown. Risk factors for the progression of DLE to SLE are difficult to ascertain because symptoms of SLE (Systemic lupus erythematous) are more severe in children than in adults, with an aggressive clinical course characterized by multi-organ involvement, particularly of the kidney, skin, cardiovascular, musculoskeletal, and central nervous systems[1]. This report aimed to describe a case of DLE in a child to establish a diagnosis, provide appropriate management, and take into account the risk of developing SLE

Case presentation –

An 8 years old female patient presented with the complaints of reddish spots accompanied by scales with black core and burning sensation in the cheeks and nose since 3 months. Small pimples and reddish spots appeared first on face then they grew. Reddish areas thickened and appeared scaly and thicker skin became more red, which then blackened and became uncomfortable, especially if the patient goes to sunlight. On physical examination, the patient's vital signs were normal. However, the antinuclear antibody (ANA) profile showed a positive value for the ribonucleoprotein/antigen Smith's (RNP/Sm), indicating the presence of an autoimmune process, one of which caused SLE. A dermatological examination of the facial region showed partially hyper-pigmented erythematous plaques, multiple sharply defined scales, and partially confluent plaques (Figure 1). On dermoscopic examination, initial lesion was found erythematous with white scales, follicular plug, and whitish perifollicular halo on the sides of the lesion (Figure 2).

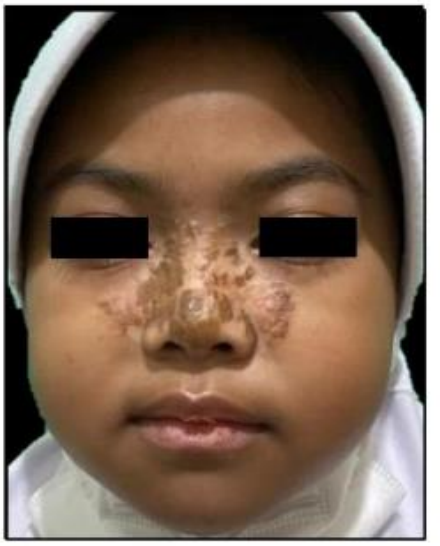

Fig.1- Clinical Appearance of Lesion on the Facial Region

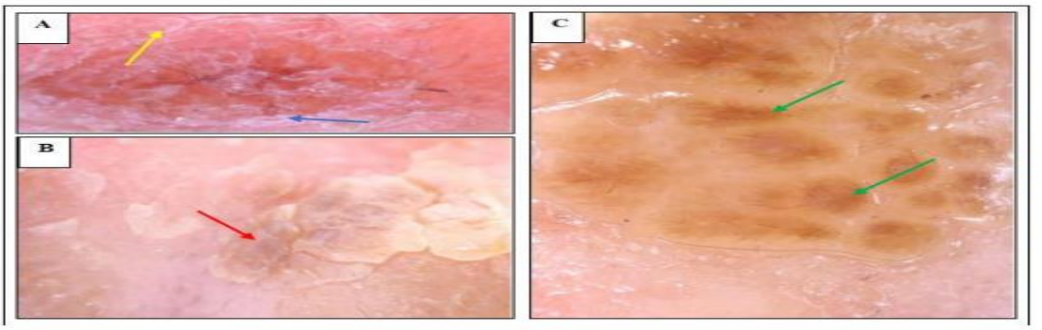

Fig.2 Dermoscopic examination of the lesion

On histopathological examination, skin tissue found to have atrophic epidermis, with follicular plug in the dermis lymphocyte and histocyte infiltration especially on perivascular and periadnexal without any signs of malignancy. Based on the above examination results support the diagnosis of DLE. Patient was diagnosed as DLE based on his anamnesis, physical examination, and histopathology. Treatment started with sunscreen, mometasone furoate 0.1% cream twice daily, and moisturizer containing glycyrrhetic acid twice daily for two weeks. Clinical improvement was seen in the form of diminished hyper-pigmented lesions in the facial region during the second week of therapy, but new lesions were found in the left buccal region. Therapy was replaced from mometasone furoate 0.1 % cream to desonide 0.05 % cream after the second week of therapy and continued with other medications. After four weeks of therapy, old lesions in the facial region appear to increase again while the lesion in the left buccal region had improved. The patient was then prescribed with systemic corticosteroid methylprednisolone 12 mg/day and mometasone furoate 0,1% cream applied to the lesion area. After eight weeks of treatment the dose was tapered to 8mg/day, old lesions in the facial region were significantly reduced, and lesions in the left buccal regions also improved (Figure 3).

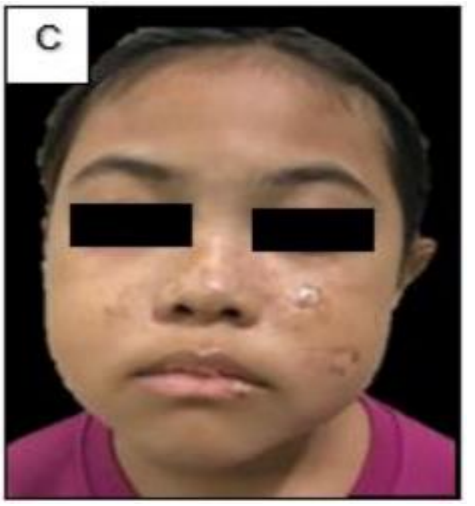

Fig.3- After eight weeks of therapy

Discussion –

Based on the patient's history, clinical examination, laboratory results, and histology, this case is identified as DLE. In this instance, the facial region displayed clinical indications of DLE in the form of erythematous plaques, fine scales, hyper-pigmented areas, and multiple discrete, partially crusted boundaries. The results of the ANA profile examination being positive point to the possibility of systemic involvement. During a dermoscopy examination, the side of the lesion exhibited erythema with white scales, a follicular plug, and a whitish perifollicular halo. A study by Ashraf et al., said that the most frequent inducing factor for DLE was ultraviolet radiation (UVR) exposure and face was the most commonly affected site. Over half of the patients had positive ANA results, and the majority of patients had localised DLE[3]. A case report by Al Janahi et al., reported a lupus erythematosus of a 3-month old female baby which upon examination found erythematous and hyper-pigmented patches and laboratory data showed positive for ANA, anti-SSA and -SSB antibodies were also positive[4].

Conclusion – This study concludes a case on DLE based on the patient's medical history dermoscopy, histology and ANA profile. Effective treatment lies in educating patient that increase the risk and selecting appropriate medications depending on symptoms.

Educating the patient on risk factors and drug selection according to complaints is the key to successful therapy for discoid lupus erythematosus.

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